VIRGINIA DEPARTMENT OF HEALTH
Protecting You and Your Environment

Protocols for Diagnostic Audiological Assessment
Virginia Early Hearing Detection and Intervention Program
Virginia Department of Health

This document provides guidance and recommended procedures for how best to implement audiological services requirements that are specified in the *Code of Virginia*, Section 32.1-46[1] and *Regulations for the Administration of the Virginia Hearing Impairment Identification and Monitoring System*[2].

The audiological assessment protocols were first developed in 1999 and revised in 2004. The 2011 revision is the product of a Virginia Department of Health (VDH) Virginia Early Hearing Detection and Intervention Program (VEHDIP) Task Force comprised of six audiological experts with extensive pediatric experience. The VEHDIP Task Force and the VEHDIP Advisory Committee— which consists of representatives from relevant groups including, but not limited to, physicians, otolaryngologists, audiologists, speech pathologists, nurses, and parents—unanimously agreed that Virginia diagnostic audiological protocol standards have followed, and should continue to follow, an exceptional model of evidence-based practice and should reflect an excellence beyond minimal standards of care. This document reflects that philosophy.

It is important to recognize that newborn hearing screening is only one component of a comprehensive approach to the management of childhood hearing loss. The process also requires follow-up diagnostic services, counseling, intervention programs, and parental education. This comprehensive process should involve a multidisciplinary team including, but not limited to, audiologists, physicians, educators, speech/language pathologists, nurses, and parents. This document, therefore, highlights key information about which audiological staff should be aware.

VEHDIP goals are to identify congenital hearing loss by 3 months of age following the Centers for Disease Control and Prevention *1-3-6 methodology*:

1 – All newborns will be screened for hearing loss **before 1 month** of age.

3 – All newborns who have failed their hearing screen will receive a diagnostic evaluation **before 3 months** of age.

6 – All infants diagnosed with hearing loss will be enrolled in early intervention services **before 6 months** of age.

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[1] To access the *Code of Virginia* citation, go to: http://leg1.state.va.us/cgi-bin/legp504.exe?000+cod+32.1-64.1
[2] To access the *Regulations for the Administration of the Virginia Hearing Impairment Identification and Monitoring System,* go to http://leg1.state.va.us/000/reg/TOC12005.HTM#C0080

All infants must be given a hearing screen prior to hospital discharge (per Regulation). The hospital discharging the infant to home should screen the infant's hearing. Even if the infant was screened and passed at a previous facility, the discharge hospital should perform a hearing screen, as the infant's health status may have changed.

I. Audiologist and Facility Qualifications

Quality audiological assessment is the diagnosis of normal hearing or hearing loss specific to each ear. The assessment of hearing loss includes identification of type and severity and, whenever possible, the nature of the hearing loss as well as knowledge of the choice of treatment options.

While any licensed audiologist may perform a hearing screen, VDH maintains a roster of audiology facilities approved for screening and diagnostic services under VEHDIP. VDH-approved audiological facilities must have the appropriate diagnostic equipment to perform **all** the specific recommended diagnostic protocols. In addition, only a licensed audiologist may perform the assessments in an approved facility. Furthermore, the approved facility must agree to the terms and conditions listed on the "Practitioner/ Facility Registration" form, which includes submitting a report to VDH within two weeks of screening. A list of approved sites is located on the VEHDIP website www.vahealth.org/hearing. The list of approved facilities is communicated to all Virginia birthing hospitals and is included with every letter VDH sends to parents. The application for approval as a diagnostic site can be downloaded from the VEHDIP website (noted above) and submitted to VDH as indicated.

II. Interpretation of Hearing Screen Results

Definitions of hearing screening results categories and the recommended subsequent actions are listed below.

- Missed: Infant not born in a hospital or not screened at the hospital prior to discharge. Screening should be performed before **1 month** of age.
- Fail: Infant failed the initial screen. Re-screening should be performed within one month of the failed initial screen **and/or** a comprehensive diagnostic evaluation by 3 months of age. Repeat screenings should not delay diagnostic evaluation in children that fail an initial screen.
- Pass with risk: Infant passed the initial screen, but one or more risk indicators for developing hearing loss later have been identified. A diagnostic audiological assessment should be completed by 24 months of age.

  Note: For re-screening, a complete screening on both ears is recommended, even if only one ear failed the initial screen.

III. Screening and Assessment

Persons providing audiological services to infants after hospital discharge:

- Must provide the screening or evaluation results to the parent or guardian (per Regulation), and should provide this information at the time of the visit.

- Must provide the child's Primary Healthcare Provider (PHP) with screening results (per Regulation).
- Should provide referral to Part C Early Intervention and/or other educational programs.
- Must give resource information to the parent [or guardian] of any child found to have a hearing loss, including but not limited to, the degrees and effects of hearing loss, communication options, amplification options, the importance of medical follow-up, and agencies and organizations that provide services to children with hearing loss and their families (per Regulation).
- Must send the Audiological Reporting Form—including test results, diagnosis, and recommendations—to VEHDIP within two weeks of the patient visit (per Regulation). The VDH Audiological Reporting Form, including risk indicators and instructions, can be downloaded from the VEHDIP website www.vahealth.org/hearing.

Outlined below are descriptions and procedures regarding initial screening, re-screening, diagnostic visit, additional assessment considerations, test parameters and thresholds, confirmed hearing loss, unilateral hearing loss, and auditory neuropathy spectrum disorder.

A. Initial Screening

A variety of technologies are available to identify hearing loss in the first days of life. The two methodologies generally accepted as effective for universal newborn hearing screening are:

1) **Auditory brainstem response (ABR)** – reflects the activity of the cochlea, auditory nerve, and auditory brainstem pathways.

2) **Otoacoustic emissions (OAE)** – reflects sensitivity to outer hair cell dysfunction.

These techniques are physiological measures of the status of the peripheral auditory system that are highly correlated with hearing status. The techniques permit the identification of infants with communicatively significant hearing impairment without referring large numbers of normally hearing infants for unnecessary follow-up testing.

Due to the increased incidence of auditory neuropathy in the neonatal intensive care unit (NICU) patient population, newborns who receive this level of care for more than five days should have both ears screened using ABR testing prior to hospital discharge or transfer to a lower level of newborn services.

B. Re-screen (within 1 month of hospital discharge)

All Cases:

- Infants who fail an OAE for their initial screening should be re-screened with an ABR.
- Infants who fail an ABR for their initial screening should not be re-screened with an OAE.

- Evaluation of both ears is necessary, even if only one ear failed the initial screening.
- Obtain all information requested on the VEHDIP Audiological Reporting Form; this information is crucial for tracking purposes.
- Obtain parent or guardian report of observed behavioral responses to auditory stimuli.
- Perform an otoscopic examination.
- Obtain and record specific results for each ear.

OAE Pass or Pass with Risk:

- If the infant passes (three of four frequencies tested), the evaluation is complete unless the audiologist determines a need for comprehensive evaluation based on medical history.
- The family should receive information about hearing, speech and language milestones, and information regarding risk indicators if present.
- For infants who pass but are at risk for progressive or delayed-onset hearing loss, the parent or guardian should be counseled regarding the need for audiological follow-up and continued monitoring.

OAE Fail ("Fail" in one or both ears, or OAE is normal but Auditory Neuropathy Spectrum Disorder (ANSD) is suspected:

- Evaluate using ABR to high-level click stimuli (80-90 dBnHL), tested with positive and negative polarity clicks in separate trials, through insert earphones.
- A trial run with the sound-delivery tube clamped should be used to differentiate between the cochlear microphonic (CM) and stimulus artifact.
- If facility is unable to perform ABR at same visit, schedule diagnostic ABR at VDH-approved diagnostic facility within one month from date of hospital discharge.

C. Diagnostic Visit (minimal requirements for audiologic 3 months assessment)

- Perform an otoscopic examination.
- Evaluate using ABR with threshold click ABR (25-30 dBnHL or lower) for each ear.
- Bone conduction threshold if click stimuli are elevated (25-40 dBHL).
- Ear-specific bone conduction if thresholds are elevated bilateral and/or an asymmetric loss is evident.
- Threshold tone burst to 500 Hz, as well as 3000 Hz or 4000 Hz.
- If facility has technology available to perform frequency-specific evoked potential testing, this may enhance diagnostic and (re)habilitative services for the infant.
- Repeat OAEs to again evaluate cochlear function and rule out ANSD.
- Counsel parents or guardians as to results and follow-up recommendations.

D. Additional Assessment Considerations

- Acoustic immittance measures using high frequency (660 or 800 Hz) tone if infant is less than 4 months of age. A 226 Hz probe tone may be used with reliability for infants 4 months or older.
- Acoustic Reflex thresholds at 500 Hz, 1000 Hz, and 4000 Hz.
- Behavioral observation to speech stimulus, 500 and 2000 Hz (minimally).
- Visual reinforcement audiometry can be effectively used for infants 6 months of age or older as a component of the evaluation. If the child will tolerate them, insertion earphones may be utilized for ear-specific information.
- Warbled pure tone or narrow band noise in sound field. Identify any minimal responses and attempt to obtain startle response.

E. Test Parameters/Thresholds (Adapted from *Stapells and Oats, 1997*)

Transient Evoked Otoacoustic Emissions (TEOAE):
Stimulus: air conduction click
Intensity: 80 +/- 3 dB SPL
Pass Criteria:

- Frequencies 2000 Hz through 5000Hz
- Three of four frequencies having reproducibility minimally: 70% @ 2400, 3200, 4000 and 5000 Hz

Distortion Product Otoacoustic Emissions (DPOAE)
Stimulus: pure tone complex
Intensity: maximum levels <70 dB SPL
Pass Criteria:

- F2 = 2000, 3000, 4000 and 5000 Hz
- Three of four frequencies have a distortion product (2F1-F2) amplitude ≥6dB than measured noise floor levels

Auditory Brainstem Response (ABR)
Stimulus: air conduction click stimulus for both ears
Pass Criteria: replicable wave V response thresholds ≤ 25-30 dBnHL

ABR Bone Conduction Testing
Stimulus and recording parameters:

- Alternating Click, 21.1/sec
- Window 15 msec
- Low Filter 30
- High Filter 1500
- Gain 100,000
- Earlobe/mastoid non-inverting electrode

Bone Conduction 2-Channel Recording (Ipsilateral/Contralateral)
Setup:

- Right-Ref I
- Left-Ref II
- High Forehead - Active jump between Ch I & Ch II

- Low Forehead – Ground

Bone Conduction Stimulus: Ear-specific 2-Channel Recording

- At higher intensity levels, evaluate ipsilateral vs. contralateral recording.
- If there is a Wave I on the ipsilateral in conjunction with the absence of a Wave I on the contralateral ear, then it can be assumed that the response is being generated from the stimulated ear.
- Record Wave V responses down to threshold.

Recording Parameter Guidelines for Tone Burst ABR

Air Conduction

EEG Channels:

- Vertex-left mastoid (Cz-M1)
- Vertex-right mastoid (Cz-M2)

EEG filter:

- High Pass: 20-30 Hz
- Low Pass: 1500-3000 Hz

One-channel recordings (Cz-ipsilateral mastoid)

- (Slope: 6 or 12 dB/octave, analog)

Window: 25 msec

Polarity: Alternating onset (compare condensation and rarefaction clicks if ABR is absent to detect CM).

Rate: 39.1/s

Ipsilateral Noise: Band-reject (notched noise, with 1-octave-wide notch centered on the tone frequency)

Contralateral Noise: White noise 30dB below tone masking level (when possible)

Stimuli 2-1-2 linear:

- 500 Hz: 4ms rise/fall, 2 ms plateau
- 1000 Hz: 2 ms rise/fall, 1 ms plateau
- 2000 Hz 1 ms rise/fall, 0.5 ms plateau
- 4000 Hz: 0.5 rise/fall, 0.25 ms plateau

F. <u>If hearing loss is confirmed</u>:

- Notify the child's PHP.
- Refer to an otolaryngologist for otologic evaluation and to obtain medical clearance for amplification if appropriate.
- Refer for amplification and assistive technology; if appropriate, provide information on cochlear implant.
- Refer children with hearing loss for genetic testing and counseling.
- Refer to Part C Early Intervention services, regardless of type or severity of the loss.
- Provide parent or guardian with information about services needed for specific type and severity of hearing loss.

G. Unilateral Hearing Loss

Unilateral hearing loss refers to a hearing loss in one ear—whether mild or profound, congenital or acquired—that warrants a definitive diagnosis. Parents or guardians have the right to know what their child's hearing level is in each ear. Depending on the etiology of the unilateral loss, there may be associated medical conditions that require further medical attention and management.

Children with a unilateral hearing loss are at risk for progressive and/or bilateral hearing loss. Unilateral hearing loss has significant implications for a child's development, socialization, and success in school. A child with unilateral hearing loss:

- May exhibit delays in speech and language development.
- Will have difficulty localizing sounds; therefore, crossing the street or riding a bike are safety concerns.
- May exhibit behavior and/or social problems.
- May have difficulty following directions, be distractible, and may appear inattentive; may show signs of fatigue as the school day progresses.
- May lag substantially behind in math, language, cognitive and social functioning as compared to their peers, and may be at risk for failing a grade in school.
- May benefit from assistive listening devices in the classroom.

Children with a unilateral hearing loss should receive:

- Routine audiologic evaluations at six-month intervals to monitor for progressive or delayed-onset hearing loss.
- Referral to Part C Early Intervention services for developmental evaluation and support. Visit www.infantva.org for information on referral processes and contacts.
- Noise protection counseling to protect the "good ear."
- Prompt medical management of middle ear disorders.

H. Auditory Neuropathy Spectrum Disorder (ANSD)

According to the Joint Committee on Infant Hearing, a small population of infants, particularly those cared for in the NICU, may be at increased risk for neural conduction and/or auditory brainstem dysfunction, including ANSD. This is a recently identified disorder characterized by a unique constellation of behavioral and physiologic auditory test results. Behaviorally, children with ANSD have been reported to exhibit mild to profound hearing loss and poor speech perception. Physiologic measures of auditory function demonstrate the finding of normal otoacoustic emissions and intact cochlear microphonic (CM) suggesting normal outer hair cell function and atypical or absent auditory brainstem response suggesting neural conduction dysfunction. CMs will show a characteristic reversal in polarity with reversal in polarity of the stimulating click; ABR will show a constant polarity regardless of polarity of the click. CMs generally remain present in individuals with ANSD despite loss of OAEs.

ABR recordings might appear as (1) a "flat" ABR with no evidence of any peaks, (2) presence of early peaks (waves up to III) with absence of later waves, or (3) some

poorly synchronized but evident later peaks (wave V) that appear only to stimuli at elevated levels.

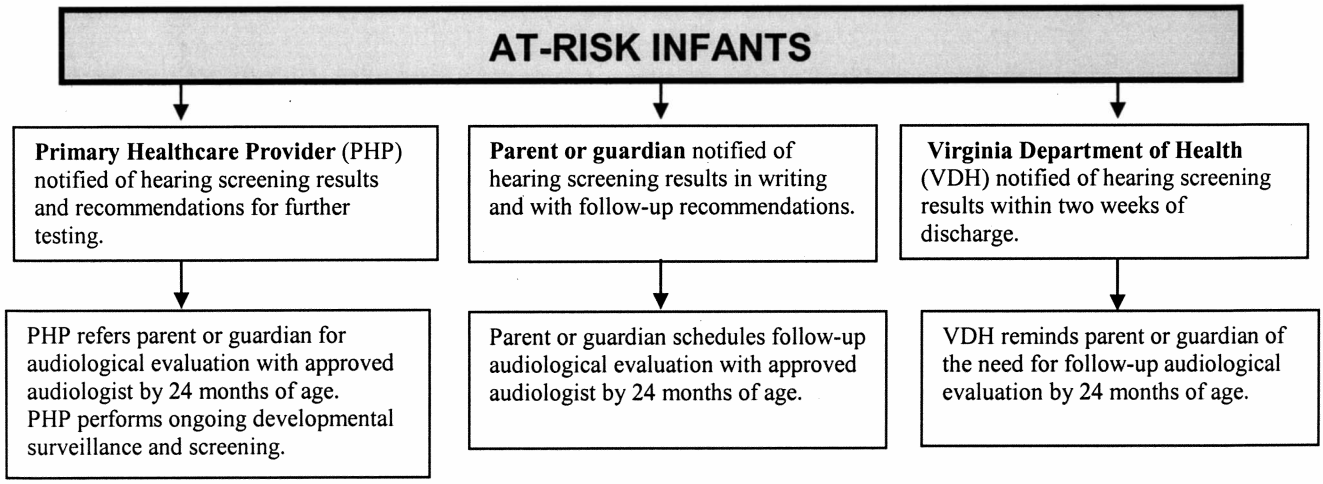
Reports suggest that those at increased risk for ANSD are infants with a compromised neonatal course, infants with hyperbilirubinemia, and children with a family history of childhood hearing loss. Currently, neither the prevalence of ANSD in newborns nor the natural history of the disorder is known. Audiologic and medical monitoring of infants at risk for ANSD is recommended by 24 months of age.
(Adapted from *Guidelines for Identification and Management of Infants and Young Children with Auditory Neuropathy Spectrum Disorder*, NHS 2008)

V. Risk Indicators

The *Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs, Joint Committee on Infant Hearing* identifies risk indicators that often are associated with infant and childhood hearing loss (see Table I). These indicators place an infant at risk for progressive or delayed-onset sensorineural and/or conductive hearing loss.

Some indicators may not be determined during the course of the hospital stay. Therefore, infants and young children who have late-onset or late-identified risk indicators should be monitored for speech, language, and hearing developmental milestones by the PHP during well-child visits.

The following diagram summarizes processes that should be followed to ensure infants and children who are at risk for hearing loss receive appropriate audiological follow-up



VDH recommends that a medical professional obtain the risk information from the infant's and mother's charts; family history of permanent childhood hearing loss should be identified by a direct question to the parent(s). The parent should not simply be given the whole list of indicators to check off, as they may not know about or understand the meaning of all indicators. The medical professional should review the risk information with the parent.

Some of these indicators are not present and/or would not be identified in the newborn period. These include parental concern and some neurodegenerative disorders or sensory motor neuropathies. These are included in the risk indicator list because parents and physicians should be informed about all indicators that can contribute to development of hearing loss beyond the newborn period.

Infants who pass but have an identified risk indicator for progressive or delayed-onset hearing loss (**pass with risk**) should have a complete diagnostic evaluation by 24 months of age.

VI. Contacts

For more information or further assistance, families are encouraged to contact:

Virginia Department of Health
Office of Family Health Services
Virginia Early Hearing Detection and Intervention Program
109 Governor Street, 8th Floor
Richmond, Virginia 23219
Phone: Toll Free 1-866-493-1090 TTY 7-1-1
Fax: 804-864-7721
Website: **www.vahealth.org/hearing**

Table I. Risk Indicators for Progressive or Delayed-Onset Hearing Loss
(For Use with Neonates and Infants Through 2 Years of Age)

Family history of permanent childhood hearing loss		
Mother of child	Grandmother of child	1st cousin of child
Father of child	Grandfather of child	More than one relative of the same parent
Sister of child	Aunt of child	
Brother of child	Uncle of child	
Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or Eustachian tube dysfunction		
• Branchio-oto-renal (BOR)	• Stickler	• Trisomy 21 – Down syndrome
• Noonan	• Williams	• Trisomy 18 – Edwards syndrome
• CHARGE association	• Zellweger	• Trisomy 13 – Patau syndrome
• Pierre Robin	• Goldenhar (oculo-auriculo-vertebral or OAV)	• Trisomy 9 – Mosaic syndrome
• Rubenstein-Taybi	• Trisomy 8 – Warkany syndrome	
Postnatal infections associated with sensorineural hearing loss		
• Confirmed bacterial meningitis	• Confirmed viral meningitis	
In utero infections		
• Cytomegalovirus	• Rubella	• Toxoplasmosis
• Herpes	• Syphilis	
Neonatal indicators		
• Intensive care greater than (>) 5 days	• Exposure to ototoxic medications: at risk aminoglycoside exposure	• Hyperbilirubinemia requiring exchange transfusion
• Extracorporeal membrane oxygenation (ECMO)	• Mechanical ventilation	
Syndromes associated with progressive hearing loss		
• Neurofibromatosis	• Jervell & Lange-Nielson	• Usher
• Osteopetrosis	• Waardenburg	
• Alport	• Pendred	
Neurodegenerative disorders, such as		
• Hunter syndrome	• Charcot-Marie-Tooth syndrome	• Friedreich's ataxia
Head trauma requiring hospitalization		
• Basal skull/temporal bone fracture	Other – specify if chosen	
Parental or caregiver concern regarding hearing, speech, language, and or developmental delay		
Craniofacial Anomalies		
• Pinna	• Atresia	• Choanal atresia
• Cleft palate	• Microtia	• Temporal bone anomalies
Chemotherapy		

Based on *Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs, Joint Committee on Infant Hearing*

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Approved by:

Karen Remley, MD, MBA, FAAP
State Health Commissioner

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